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THE OPERATIVE TREATMENT OF SPINA BIFIDA AND HYDROCEPHALUS, BY JAMES H. NICOLL, M.B.

THE OPERATIVE TREATMENT OF SPINA BIFIDA AND HYDROCEPHALUS.

By JAMES H. NICOLL, M.B.,

Assistant Surgeon, Western Infirmary; Extra Surgeon, Children's Hospital; Consulting Surgeon, Ear Hospital; Consulting Surgeon, Glasgow District Lunacy Board, etc.

A YEAR ago I read before the section of Diseases of Children at the meeting of the British Medical Association in Edinburgh a paper on the "Radical Cure of Spina Bifida," based on an experience of thirty-two cases which I had treated up to that time. That paper was published in the *British Medical Journal* for October 15th, 1898. It was subsequently, on the request of the editor of that journal, published in *Pediatrics*, and reproduced in brief in the *Archives of Pediatry*, and it has since been quoted by several writers.

During the year which has intervened since the paper was published, I have had a number of letters from professional brethren requesting advice on cases of Spina Bifida and Hydrocephalus, and have had referred to me twenty-one further cases of these affections. Of these cases several presented features of sufficiently exceptional interest to warrant their inclusion in such a publication as the *Glasgow Hospital Reports*. And, in view of the publicity which has been given to my latest paper, it may be well to take this opportunity of summarizing what I have at various times published on the subject in the *Glasgow Medical Journal*, the *British Medical Journal*, and the transactions of several societies.

The conclusions which the results so far have led me to may be stated as follows:

That treatment by injection, whatever its merits in times past, is, in comparison with the open operations which modern aseptic practice has made possible, uncertain in its results, of high mortality, and unsurgical in conception.

That open operation, in suitable cases, has a mortality little, if at all, higher than has the operation for the radical cure of inguinal hernia. That this does not apply to cases where, on account of difficulty over nerve tissues, incomplete or partial operations have to be adopted. That the term "suitable case" applies to cases in which the sac is not ruptured, ulcerating, or sloughing, and in which there is not present an extreme degree of hydrocephalus. The presence of such conditions (*vide British Medical Journal*, October 15th, 1898), while not constituting a bar to operation, rather, perhaps, rendering the need for operation the more urgent, greatly increases the chances of the operation failing to save the life of the child.

That open operation in cases in which the conditions permit of a complete operation accomplishes a "radical cure" of spina bifida. Out of forty-six cases operated on, I have so far had only three cases in which, after complete operation, recurrence took place. Two of these were subsequently remedied by second operation.

That the radical cure of spina bifida proves in some cases a remedy for the accompanying paralysis of lower limbs, bladder, and rectum. In several cases which I have had the removal of bands of spinal cord or nerves from positions of tension, stretched over the walls of the sac, and the replacing of them in the spinal canal, have resulted in relief of the paralysis—partial in some cases, complete in others.

That in cases where hydrocephalus accompanies spina bifida the operative measures adopted for the latter may be made the means of treating the former, or combined with measures to that end. I have treated a number of cases of hydrocephalus by one or other of the methods described below. What opinion is to be formed of the

PLATE I.

(Plate IV. in publication quoted.)

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ultimate results can only be determined after a period of years. At present these cases are under observation, and in some of them a marked relief has followed treatment, while in several, cure appears to have resulted. In all cases, however, I should wish to wait until puberty is past before expressing too definite an opinion.

The methods of operating I have followed have been fully described elsewhere (*British Medical Journal*, October 15th, 1898). They have, for the most part, included the use of flaps of skin dissected from the base of the tumour, with, in addition in many cases, flaps of mesoblastic tissues (fascia, muscle, and even bone) raised from the margins of the gap in the spinal canal.

They vary chiefly in the manner of dealing with the sac of spinal membranes, and may be briefly stated as follows:

(*a*) Pure meningoceles, without contained nerve cords, are simply cut away, and the neck closed by either ligature or suture, and covered by the flaps.

(*b*) Meningoceles which contain nerve tissue—either spinal cord or spinal nerves: in such cases I have employed two different methods. One of these would appear to be in some respects new. Since I first published it it has been quoted with my name attached (*vide* the recently published text-book of *Surgery* by Pick). That being so, it may be well to reproduce here the last published account of it. The following extract, with two accompanying photographs, is taken from the paper of October 15th, in the *British Medical Journal*:

"*Cases in which the sac on being freed and opened is found to contain nerve cords.*

"In my earlier cases, several of which were shown on different dates to the Glasgow Medico-Chirurgical Society, I followed the practice of dissecting the cords off the interior of the sac This is not free from risk, and in one case, operated on in 1895, I produced damage in detaching the

nerves which caused paralysis of one leg, which has remained permanent.

"To avoid such risk I adopted a plan which, in connection with the demonstration of several cases of cured spina bifida, I described to the Glasgow Medico-Chirurgical Society in November, 1897. The method is as follows:

"Flaps of skin and mesoblastic tissues are dissected up in the usual way. The sac is freed and opened. Such portions of it as are free from nerve tissue are excised. The remainder is 'cut into ribbons'—by incisions made from the interior, parallel with the nerve cords incorporated with it—and thoroughly roughened with the point of knife. The 'slashed' and roughened sac is then placed in the patent spinal canal, over which the flaps are sutured in the usual way.

"In a certain number of cases—two of which have been mentioned above, and two of which (Plates IV. and V.) are described below—I have still further extended the principle and considerably modified the method of this operation, with good results.

"Plate IV.: M. D., shown to the Glasgow Medico-Chirurgical Society in April, 1898, was sent to me, at the age of 5, in 1896, by Dr. Lamont, of Chryston.

"*Particulars of case.*—Spina bifida tumour 8 by 9 inches across. Complete paraplegia: constant incontinence of urine: condition of bowel in which obstinate constipation alternated with incontinence of faeces and flatus.

"At that time I operated—opened sac by incision shown in plate—found it so occupied by expanded nerve cords that excision of even small portions appeared impossible. With the knife I carefully tore such small areas as seemed free from nerve tissue, and gently, but pretty thoroughly, roughened the whole interior, including the surface of the nerve cords in many parts, and stitched up.

"Condition when shown in April, 1898, nearly two years after operation:—Sac replaced by a tough lobulated fibrous mass, shown in plate. This is considerably smaller than the pre-existing sac, measuring 4 inches by 3½ inches

PLATE II.

(Plate V. in publication quoted.)

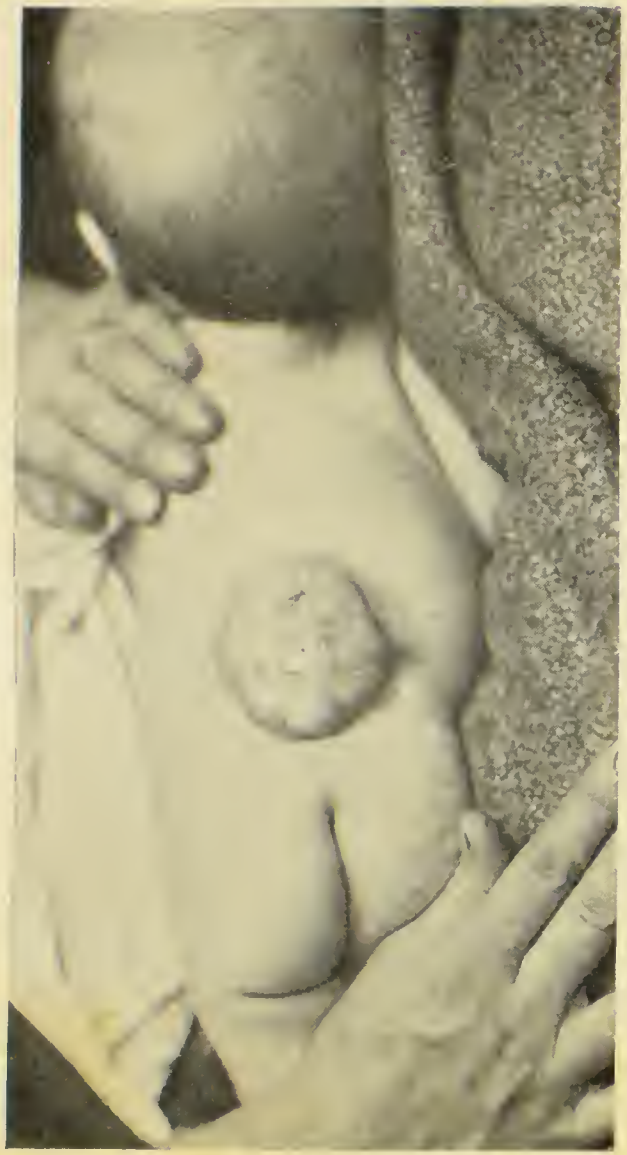

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PLATE III.

(Plate II. in publication quoted.)

EXCISION OF SPINA BIFIDA SAC.

Scar of wound eight weeks after operation, showing puckered elevated ridge produced by method of suturing. Ridge already largely obliterated.

across. It is not tender, is not altered in size by pressure, and was compared in consistence to a lipoma or a fibroma by various members of the Society who examined it. The child can walk several miles at a time, and has nearly perfect sensation in the legs, which are, however, in some respects, like the limbs of infantile paralysis, being bluish and distinctly deficient in muscular bulk. The bowel acts regularly, and the sphincter is now efficient. Urinary incontinence has completely ceased except that, as the mother states, a little involuntary escape is apt to occur if the child is made nervous or excited.

"Plate V.: Infant at present in the M'Alpin Nursing Home. The plate shows a hard fibrous mass, unaffected by pressure, occupying the site of a former spina bifida tumour of nearly thrice its size.

"The sac was opened five weeks ago by a median incision, and was found largely covered by nerve cords. Its interior was roughened in the manner above described, and closed by suture.

"It is now practically obliterated. My reason for breaking the rule I have laid down for myself, and producing this case at this early date after operation, is that, as will be gathered from the plate, the case illustrates what I believe to be a fact —viz., that, unless present in an extreme degree, hydrocephalus constitutes no bar to successful operation on spina bifida. The plate indicates what is the fact, that the patient suffers from very pronounced hydrocephalus.

"At the meeting in November, 1897, when I first described this method of operating, I stated that, in searching the literature of the subject, I had found a paper by Mayo Robson, of Leeds, in the *Annals of Surgery* for July, 1895, in which most interesting communication Professor Robson says:—'In cases where the cord is expanded, or nerves are blended with the sac, excision of redundant parts or incisions between manifest portions of nervous structures reduces the tumour, and enables it to be placed in the canal, and usually there is no difficulty in covering the replaced structures.'

"The method, therefore, into which my experience has

gradually led me would seem to be in some respects a combination of the principles of Professor Mayo Robson's operation for spina bifida and Professor Macewen's operation for aneurism.

"It differs from Professor Robson's in that no attempt is made to reduce the bulk of the tumour by the operation, that end being attained by the fibrous contraction and consolidation which follow: and, further, in that operative procedures are not confined to the portions of the sac between the nerve elements; and, finally, that in certain cases (viz., such cases as Plate I., where the sac was torn and roughened internally by a needle passed into it through the skin) the procedure has no similarity whatever with Professor Robson's operation."

Since the foregoing was published I have for certain cases (and certain cases only) adopted a modification of the operation. The "slashed" and roughened sac is gently packed with sterilized iodoform or other gauze and partially sutured. At the end of twenty-four to forty-eight hours the packing is removed and the operation completed by suture of the flaps in the usual way. It is perhaps not necessary to remark that such a method of operating is, equally with such drainage as was carried out in the case of Baby B. (*Multilocular lateral spinal cyst*), given below, only justifiable under a rigid system of special nursing. The child lies prone on a pillow on the nurse's knees for a week or longer. During that time the back is constantly exposed to view so that contamination of the dressing by urine or faeces is impossible.

It will be evident that the local result of such an operation must be less satisfactory than the result in cases where the conditions permit of a complete excision of the sac, as in Photographs II. and III. reproduced from the same paper.

In the treatment of hydrocephalus I have employed several methods described in the paper quoted. Of their comparative values I have not yet been able to form an

PLATE IV.

(Plate III. in publication quoted.)

Excision of Spina Bifida Sac.

Scar of wound three years after operation—position of extremities indicated by pencil marks. Puckering completely obliterated and scar very faintly seen.

Facing p. 302.

opinion sufficiently decided for publication. They are, briefly, the following:

(*a*) Drainage of cerebrospinal fluid at the time of operating for spina bifida. Into the vexed question of the route or routes by which hydrocephalic fluid reaches the sac of a spina bifida it is not necessary to enter here. It is sufficient to re-state the fact that in certain cases of spina bifida with hydrocephalus it is possible, by raising the child's head and shoulders after the spina bifida sac has been opened, to drain away as much fluid as may be deemed safe, the amount being judged by the tension of the fontanelle.

(*b*) Continuous drainage by a tube inserted into the spinal canal through the neck of an excised spina bifida sac at the time of operation. In several cases I have kept such a tube draining fluid into the dressings for a week.

(*c*) The establishing of a drainage channel from the spinal canal (at the site of, and during the operation for, spina bifida) into either the peritoneal cavity or the cellular subcutaneous tissue or both. This may be done by a drain which is absorbable after a time, or by a drain which is removed by secondary operation after a period of weeks or months.

(*d*) Drainage of the cerebral ventricles, either externally or into the meninges.

(*e*) Application of various solutions (chiefly iodine) to the interior of the cerebral ventricles. What experience I have had of this method has impressed me favourably. It is not, however, devoid of risk. The amount and the strength of an injection will depend on the degree of dilution likely to occur internally, that is, on what the operator knows, from previous drainage or otherwise, of the amount of fluid present. In any case, for a first injection something less potent than Morton's fluid should be employed.

Of the cases of spina bifida and hydrocephalus I have seen during the past year, some twenty-one in number, those following presented features somewhat out of the usual:

Case treated throughout as an out-patient.

In the paper in the *British Medical Journal* I stated, "It may fairly be said that in cases of spina bifida situated in the lower lumbar region the success of the operation is as much in the nurse's as in the surgeon's hands. The operation itself in cases of pure meningocele is of the simplest: and, when the tumour was placed in the dorsal or cervical region I have more than once operated on cases in the out-patient department, and had them taken home and nursed by the mothers with complete success." This case proves that spina bifida sacs situated in the lumbar region may be similarly treated.

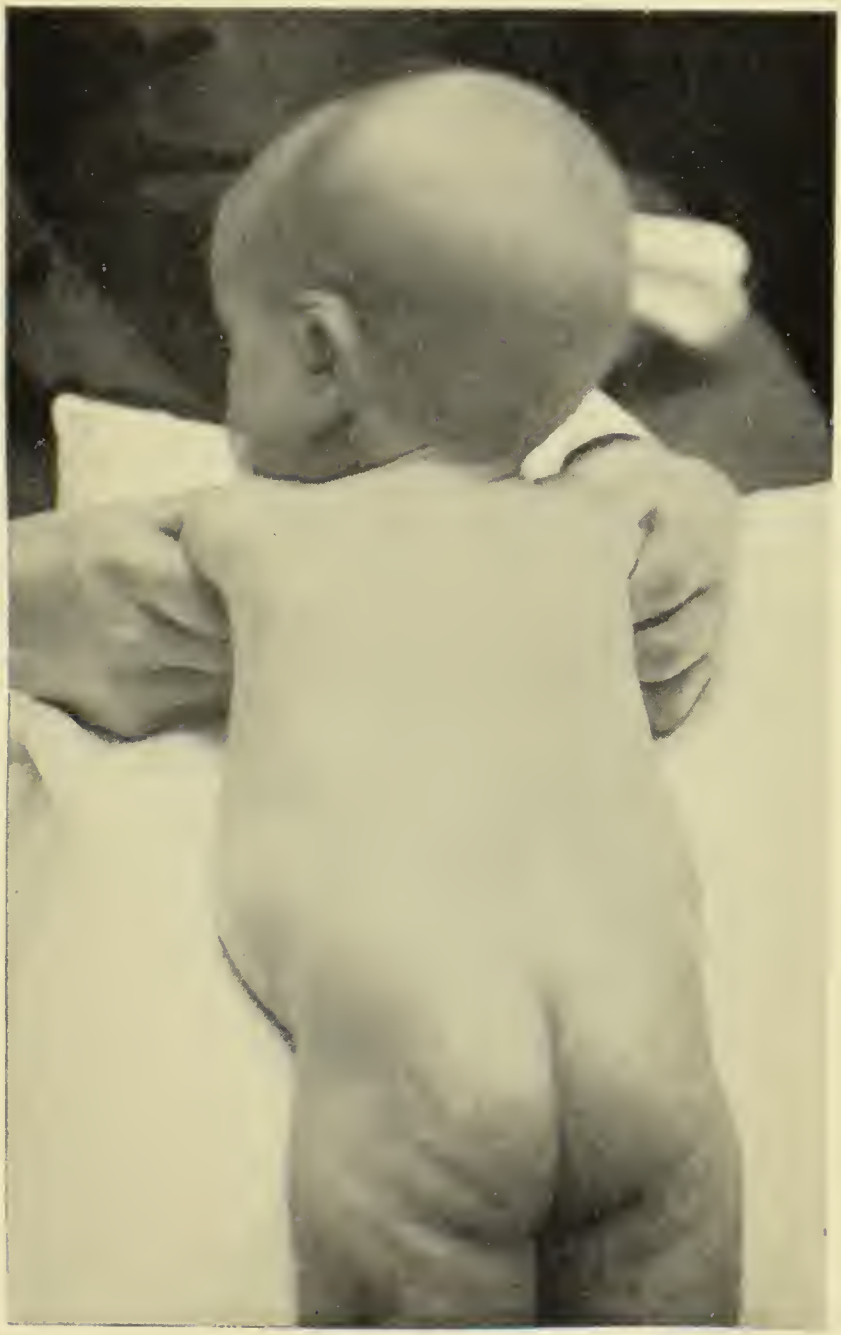
Baby D., at the age of two months, was sent to me at the Children's Hospital on January 6th last by Dr. J. Scanlan. It had a spina bifida sac in the lower lumbar region about half as large again as a duck's egg. This I excised. Sister Laura undertook the dressing, instructed the mother as to the nursing, and visited the child regularly. The result is as the photograph shows (Plate V.). Union occurred so perfectly that six months later, the date of the photograph, the scar of the operation was practically imperceptible. The patient was shown to the Glasgow Medico-Chirurgical Society on April 21st.

Case of multilocular lateral spinal cyst.

Baby B., aet. six months, sent to me in October last by Dr. R. Crawford. The photograph (Plate VI.) indicates the size of the cystic tumour, but, owing to the position of the child, fails to show the site clearly. The swelling occupied an area corresponding with the right half of the sacrum, right sacroiliac synchondrosis, and a large part of the right ilium. It was elastic, had a perceptible respiratory wave, and became tense on exertion. There was distinct hydrocephalus.

The child was admitted to the M'Alpin Nursing Home for operation. I made a free incision from the middle line outwards and downwards towards the great trochanter, and partially isolated the cyst from the surrounding

PLATE V.



Case of Excision of Lumbar Spina Bifida which was treated throughout as an out-patient. Photograph six months after operation.

Facing p. 304.

tissues. Finding isolation difficult, I incised the tumour and explored it internally. It consisted of a multiloculated sac, containing cerebrospinal fluid, and sending processes amongst the deep tissues. Crossing one portion of its space were four nerve cords, apparently going to form the great sciatic nerve. Just above this was a deep process of the cyst, and the finger passed into this seemed to go through the bone in the region of the sacroiliac synchondrosis into a recess extending into the retroperitoneal tissue. It being clear from the position of the nerve cords mentioned that any attempt at excision of this complicated and deeply-placed sac would involve the risk of paralysis of the limb, I abandoned operation, and closed the wound.

Three days later, the child progressing well, I removed one of the skin sutures and passed a rubber tube into the sac, securing it to the skin by suture. For a week that was allowed to drain. At the end of that time the tumour was much reduced in size, the fontanelle depressed (and pulseless, when the head was raised), the face shrunken, and the eyes "sunk" in the orbits. The tube was withdrawn, and another suture inserted. The child recovered perfectly, and remained well, and in April it was shown to the Glasgow Medico-Chirurgical Society. In July the child continued well, and the hydrocephalus was distinctly less marked. The cystic tumour, while of somewhat less bulk than formerly, and appreciably solid in the region of the scar, was unaltered. On July 3rd I punctured it with a trocar and cannula, evacuated some drams of fluid, and injected one dram of Morton's fluid. The result was severe illness of the child, marked by a period of over a week of high temperature and general twitchings; and at the present time, a month later, Dr. Crawford reports that the child is just regaining its usual health. The injection has, as yet, had little if any local effect on the cyst.

It need hardly be remarked that any such drainage as was carried out in this case is only permissible under nursing arrangements which are absolutely reliable; and I should wish to express my indebtedness to the nurses of the M'Alpin

Nursing Home, and to others, for the care with which they have nursed many of these cases. It is no light task to nurse a fretful infant night and day on its face for a fortnight, without once turning it round or permitting it to raise its shoulders, and without allowing the least soiling of the lumbo-sacral region. And when to this, in some cases, is added the presence of a constantly trickling drainage tube, the amount of flow from which is regulated from time to time by the relative heights of head and pelvis, the task becomes one of some little delicacy.

In Kirmisson's *Maladies Chirurgicales d'Origine Congénitale* is the record of a lateral cystic spinal tumour in the same region.

Cases in which the termination of the spinal cord was excised with the sac.

In spina bifida sacs situated in the lumbo-sacral region it is not uncommon to find a dimple produced by attachment to the dome of the sac of the terminal part of the spinal cord. In certain cases I have, after carefully isolating and displacing all accompanying nerves (constituting the cauda equina), excised this terminal attachment of the cord with the sac This greatly simplifies the operation. I have seen no resulting paralysis.

Case 1.—Baby A., puny and feeble, sent to me, at the age of 2 months, in May last by Dr. Jas. Rutherford, with a spina bifida in the lumbar region nearly as large as an average orange.

This I removed in the Western Infirmary. The child made a good recovery. Plate VII. is a photograph of the dome of the sac, with attached to it 1⅞ inches of the terminal central strand of the cord. Part of this has been removed and subjected to microscopical examination. It appears to consist solely of fibrous tissue.

The ellipse of sac and skin fused which is shown in the photograph represents a portion only of what was removed. After the flaps outlined by the elliptical incision had been

PLATE VI.

MULTILOCULAR LATERAL SPINAL CYST.
(Case of Baby B.)

Facing p. 306.

raised, and the sac isolated to its neck and opened, the remainder was removed in the usual way.

Plate VIII. is a photograph of the child taken six weeks after operation. The operation scar, as in the case of Baby D. (*vide* Plate V.), was so faint as to be all but imperceptible in the photograph. The negative was therefore "touched" in order to emphasize the impression. The result has not been happy, the impression given being that of a large irregular scar.

The child at present, three months after operation, is still decidedly feeble, as before operation. It has grown, however, and there is no indication of any paralysis.

Case 2.—Baby B., aet. 3 weeks, sent to me by Dr. Thomson, of Airdrie, in January, 1899, with a spina bifida in the lower lumbar region of the size of a Normandy pippin. This I excised in the usual way, and with it the terminal central strand of the spinal cord.

On the morning of the operation the child was found to be suffering from marked carbolic acid poisoning. Its urine was blackish-green when passed, and the child was markedly collapsed, and vomited frequently. In three cases I have seen similar symptoms follow the application of carbolic dressings to spina bifida sacs prior to operation; and it seems not improbable that the very thin parietes, bathed on one side by serous fluid, offer conditions under which absorption from the outside may occur with great facility. On the two former occasions on which symptoms of carbolic poisoning were present, I deferred operation. On this occasion the child appeared so ill that I thought it better to proceed with the operation in the hope that the evacuation of the contents of the sac might lead to relief of the symptoms. Unfortunately, this did not occur. The symptoms persisted, and were aggravated by increase of the vomiting and diarrhœa, with evidence of acute gastrointestinal irritation, accompanied by subnormal temperatures. The wound, in spite of the child's increasing weakness, healed before death occurred from exhaustion on the eighth day. During the period subsequent to operation there was no evidence of paralysis. The terminal

strand of the spinal cord which was removed was examined by Dr. Fullerton, who assisted at the operation, and myself, and is at present in my possession. It consists of a fine tube, with thick fibrous walls, covered by glistening endothelium. It is less than $\frac{1}{8}$ inch thick and about $1\frac{3}{4}$ inch long. It appears to be the filum terminale little, if at all, altered.

Case 3.—Baby B., sent to me, at the age of five weeks, by Dr. Lamont, with a spina bifida in the lumbar region of the size of a large Tangerine orange. This I removed in the M'Alpin Nursing Home in May, 1898, and with it a portion of the terminal strand of the cord measuring $1\frac{1}{4}$ inch. The child is at the present time well, and exhibits no evidence of paralysis. The part removed consists of nerve fibres and fibrous tissue.

Cases of spina bifida complicated by abnormal patency of the neurenteric canal.

Such cases are recorded as rarities from time to time in surgical literature. An illustration of one is included in Vol. II. of Treves's *System of Surgery*. The records of the two following cases are given for what they may be worth in the unfortunate absence of anatomical demonstration of the existence of a patent neurenteric canal.

Case 1.—Baby H. was brought, aged two days, to me at the Children's Hospital on May 21st, 1897. The infant was "shrivelled" and puny. The anus was all but quite imperforate, being represented by an aperture which just admitted an ordinary surgical probe. This aperture was situated on the summit of a prominent swelling, which occupied the perineal region and ischiorectal fossae, and formed the projecting portion of a mass which occupied the true pelvis, and could be felt above the pubis. In the lumbar region was a swelling which presented all the characters of a spina bifida which had ruptured by ulceration, and through the rupture was discharging a fluid which in odour, colour, and consistence was clearly liquid faeces.

I dilated the constricted anal aperture till it admitted

Dome of sac excised with terminal central strand (fibrous) of spinal cord attached.

Facing p. 308.

PLATE IX.

Photograph of Baby H.'s back, viewed from below, showing (1) Ulcerating spina bifida through which faecal material escaped; (2) Anal aperture, lacerated by previous dilatation; (3) Left leg pulled up under pelvis, showing wrinkles of loose skin due to emaciation.

my forefinger and gave vent to a quantity of semi-solid faeces. The child was sent home in the expectation that death would speedily ensue. In spite of its extreme feebleness, however, it lived for a week. During that time the faecal discharge from the lumbar orifice gradually ceased—though not entirely. Three days before death the photograph represented in Plate IX. was taken. The perineal swelling, which was evidently due to retention of intestinal matters, had by this time disappeared, the retained matters having found exit through the dilated anal orifice.

After considerable trouble permission was obtained for a post-mortem examination. This Dr. Primrose and I carried out. By means of free incisions we removed the entire lumbar spinal column with the sacrum in one piece, and, attached to this, all pelvic and abdominal structures in relation to it, including the rectum. That the swelling in the lumbar region was a spina bifida was evident in the deficiency of the laminae and spines. The specimen I put away in alcohol for dissection later. Most unfortunately it has been lost, having apparently disappeared in the course of removal of my household furniture to my present address.

Case 2.—On April 20th, 1899, I received from Dr. M'Lean, of Stonehouse, the following letter:

"I have a case here which will interest you. The child was born yesterday morning, and seems a healthy child, but has what I took to be a burst spina bifida in the lower lumbar region. When I was at the house to-day I was told that the faeces were 'coming by the back.'"

I arranged accommodation for the infant in the M'Alpin Nursing Home; but, as the weather was cold, it was deemed advisable to wait for a suitable day for the journey.

On April 28th Dr. M'Lean informed me that the child had died that day after a series of general convulsive seizures.

With a view to securing a post-mortem examination and a photograph, I at once communicated with Dr. M'Lean. To that end he used every endeavour. The parents, however, naturally enough, absolutely declined: and refused even to

permit me to see the body. Most unfortunately, therefore, an actual pictorial representation of the case is lacking.

At my request Dr. M'Lean put on record the facts of the case and sent his notes to me next day. These I reproduce here:

"The spina bifida occupied the lower lumbar region. Its size was that of a large hen's egg. It had originally evidently been larger: for it was ruptured, and clear fluid was oozing out, while the dome of the swelling was shrunken. At the neck of the sac was the orifice of a passage, large enough to admit my little finger, and surrounded by large red granulations. Through this passage the faeces were passed. The anus appeared normal, and the child had no other visible defect."

Case of cystic tumour (of patent neurenteric canal?).

B. T. was sent, at the age of three months, to me by Dr. R. Morton, on October 14th, 1898. Fig. X. represents the lower limbs and perineum of the child at that time. The cystic tumour depicted occupied a site between the anus below and the coccyx above. The coccyx was tilted backwards and upwards.

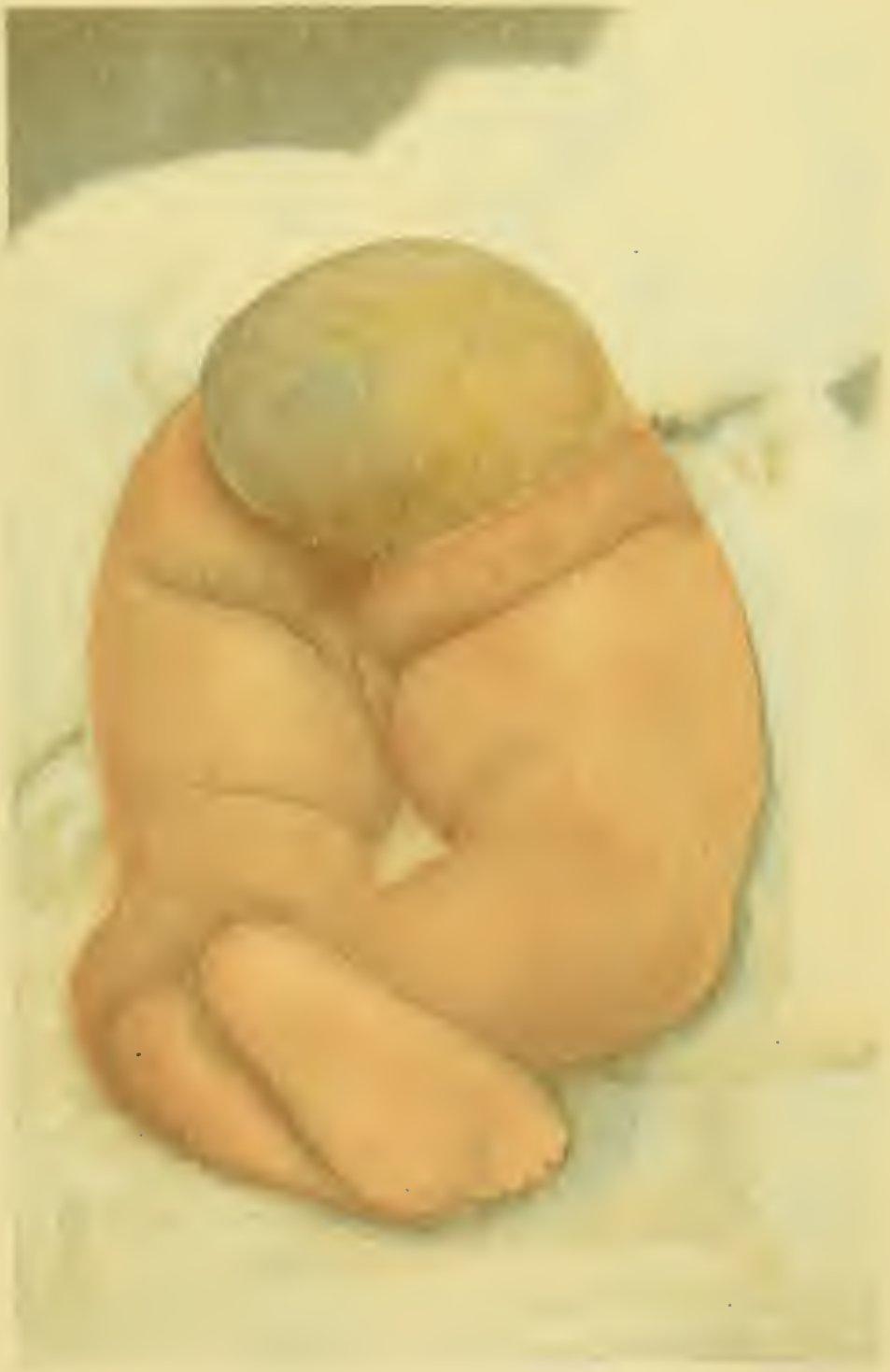
I excised the greater part of the mass. The specimen consists largely of firm fibrolipomatous tissue. It contains two cysts of the size of pigeon's eggs. One of these contained thin pultaceous material, which escaped during the operation. The other contained serous fluid. The wound healed partially, complete union being prevented by the constant escape of a clear thin fluid having all the characters of cerebrospinal fluid and the intermittent escape of bells of gas with unmistakable faecal odour.

At the end of three weeks I reopened the partially healed wound, and, dissecting more deeply, removed another cyst with thin walls and turbid serous contents. This, and the specimen formerly removed, were shown to the Glasgow Medico-Chirurgical Society on April 21st, 1899. Both are in my possession. At the end of seven weeks the wound had

PLATE VIII.

Case of Baby A.

Facing p. 310.



contracted to a fistulous aperture, and the child was dismissed from the nursing home. It was shown to the Medico-Chirurgical Society on the date mentioned. At present the child is in good health, though distinctly hydrocephalic. The fistulous tract below the coccyx persists. No gas escapes now, but there is a frequently recurring flow of thin serous fluid in small amount.

The exact nature of this case is not quite clear. My impression is that the tumour was a multilocular cyst taking origin in a partially obliterated neurenteric canal, and that the operation opened the tubular communications with both spinal canal and rectum.